British Heart Journal, 1974, 36, 1049–1060.

Haemodynamic and anatomical characteristics of pulmonary blood supply in pulmonary atresia with ventricular septal defect – including a case of persistent fifth aortic arch

F. J. Macartney, Olive Scott, and P. B. Deverall

From the Leeds Regional Cardiothoracic Unit, Killingbeck Hospital, Leeds

Eight children with congenital pulmonary atresia with ventricular septal defect were investigated. The anatomy of pulmonary blood supply was established by aortography and multiple selective injections of contrast medium into aortopulmonary anastomoses or pulmonary arteries. All patients were shown to have central pulmonary arteries (derived from the embryological sixth aortic arches). In 4 (group 1) the central pulmonary arteries had been supplied via a persistent ductus arteriosus alone (3 subsequently had surgical aortopulmonary shunts). In 3 (group 2), the central pulmonary arteries were supplied via major aortopulmonary collateral arteries alone, and in 1 patient (group 3) pulmonary blood supply was derived both from collateral arteries and a persistent left fifth aortic arch. Systolic pressure gradients of 28 to 81 mmHg were demonstrated in each patient at the pulmonary artery end of the aortopulmonary anastomosis, whatever its nature.

All patients in group 1, and 1 patient in group 2 had unifocal pulmonary blood supply, i.e. the sixth aortic arch anastomosed with, and provided a single focus for, all systemic sources of pulmonary blood supply. Pulmonary resistance relative to sixth aortic arch pressure ranged from 1·4 to 6·4 units m^2.

The remaining patients had multifocal pulmonary blood supply, i.e. one or more aortopulmonary collateral arteries provided pulmonary blood supply independent of the sixth aortic arch. In this group the minimum pulmonary resistance relative to the sixth aortic arch ranged from 3·9 to 12·5 units m^2. The latter figure suggested the presence of severe pulmonary vascular disease.

Thus stenoses at the aortopulmonary collateral/pulmonary artery junction usually, but not invariably, protect the pulmonary vascular bed from the effects of perfusion at aortic pressure.

The surgical correction of persistent truncus arteriosus and pulmonary atresia with a ventricular septal defect has been made possible by the development of valve-bearing conduits which are used to establish continuity between the right ventricle and the pulmonary circulation (Ross and Somerville, 1966; McGoon, Rastelli, and Ongley, 1968). It is now recognized that in both conditions the resistance to blood flow in the pulmonary circuit anastomosed to the conduit is a major determinant of survival after operation (McGoon *et al.*, 1968). The haemodynamics of the pulmonary circulation in truncus arteriosus are fairly well described (Tandon, Hauck, and Nadas, 1963; Victorica *et al.*, 1969), but this is not true of pulmonary atresia, largely because of the inaccessibility of the central pulmonary arteries. The need for preoperative information on this point is amply demonstrated by 2 case reports of patients with pulmonary atresia and ventricular septal defect, whose right ventricular pressure fell only slightly after operative correction. One patient died and the other has persistent pulmonary hypertension (Somerville and Ross, 1972; Doty *et al.*, 1972). The realization that it was relatively easy to enter major aortopulmonary collateral arteries with a cardiac catheter (Chesler, Beck, and Schrire, 1970; Macartney, Deverall, and Scott, 1973) prompted us to apply this technique to those cases of pulmonary atresia with a ventricular septal

Received 21 March 1974.

defect in which there are well-developed intrapericardial pulmonary arteries.[1]

Subjects and methods

The case material consists of 8 children (Table) in whom the diagnosis of congenital pulmonary atresia with a ventricular septal defect had been established by clinical examination and selective right ventricular angiocardiography early in life, before any attempt at palliative surgery had been made. Patients with single (or common) ventricle or transposition of the great arteries were excluded. In all patients the presence of confluent left and right pulmonary arteries (Edwards and McGoon, 1973) was established by rapid biplane aortography and frontal plane cineangiograms of selective injections of contrast medium into either major aortopulmonary collaterals or the intrapericardial pulmonary arteries themselves. The method used for catheterization of aortopulmonary collateral arteries has already been described (Macartney *et al.*, 1973). Patients were studied fasting and supine, premedicated with 1 ml/9 kg of a mixture containing 25 mg pethidine, 6·25 mg chlorpromazine, and 6·25 mg promethazine per ml (maximal dose 3 ml).

In pulmonary atresia with a ventricular septal defect the aortic end of the ductus arteriosus when present tends to be directed cranially rather than caudally (Rudolph, Heymann, and Spitznas, 1972; Miller *et al.*, 1973). Catheterization of a persistent ductus arteriosus (Fig. 1) was therefore very much easier from the aortic arch than from the descending aorta. The axillary artery on the side of the aortic arch was usually selected for arteriotomy because the origin of the corresponding subclavian artery is usually close to the ductus.

When a surgical anastomosis between the ascending aorta and right pulmonary artery had been created, entry into the right pulmonary artery from the aorta was achieved either retrogradely from the ascending aorta or with a transvenous catheter advanced from the right ventricle through the overriding aortic valve (Fig. 2a). Entry into the left pulmonary artery by this method was not accomplished. In the one patient (Case 3) with a Blalock-Taussig anastomosis, both pulmonary arteries were entered with a transvenous Sones catheter advanced via the aortic root and innominate artery.

Passage of a cardiac catheter through any aortopulmonary anastomosis in pulmonary atresia may result in interruption of the only source of pulmonary blood supply. Therefore, in all patients except the first investigated (Case 1), before entry to the pulmonary artery was attempted, all preparations for measurement of oxygen uptake were completed, a catheter was placed in the superior vena cava to sample mixed venous blood, and an indwelling needle placed in the femoral artery. When the pulmonary artery was entered, samples were drawn and pressures measured immediately for calculation of pulmonary and systemic flows and resistances. Systemic arterial pressure and saturation were monitored throughout the period in which the catheter lay in the pulmonary artery. Oxygen uptake was determined by the flow-

[1] The terms 'intrapericardial pulmonary arteries' and 'sixth aortic arches', though not exactly equivalent, are used interchangeably throughout this paper in an attempt to bridge the gap between 'practical' surgery and 'theoretical' embryology. It is assumed that, if an artery to the left or right lung is identified within the pericardium, it is a sixth arch derivative.

TABLE *Clinical and haemodynamic data*

Group	*Case No.*	*Age (yr)*	*Sex*	*Aortic O_2 satn (%)*	*Hb (g/100 ml)*	*Previous shunt*	*Right pulmonary artery pressure (mmHg)*	*Left pulmonary artery pressure (mmHg)*	*Aortic pressure (mmHg)*
1	1	1	F	66	18·2	—	8/2	8/2	89/46
	2	3	F	75	22·7	Aorta – right pulm. artery	12/3	12/3	98/59
	3	14	M	79	17·6	Rt. subcl. – right pulm. artery	21/4	21/4	92/52
	4	5	M	77	17·4	Aorta – right pulm. artery	8/2	—	100/48
2	5	10	F	80	12·9	Aorta – right pulm. artery	82/58 78/50	18/5 —	110/62 108/64
	6	7	F	79	14·1	—	35/24	10/0	110/55
	7	4	F	80	13·7	—	34/17	—	95/47
3	8	4	F	79	13·3	—	78/55 58/42	78/55 58/42	110/55 100/50

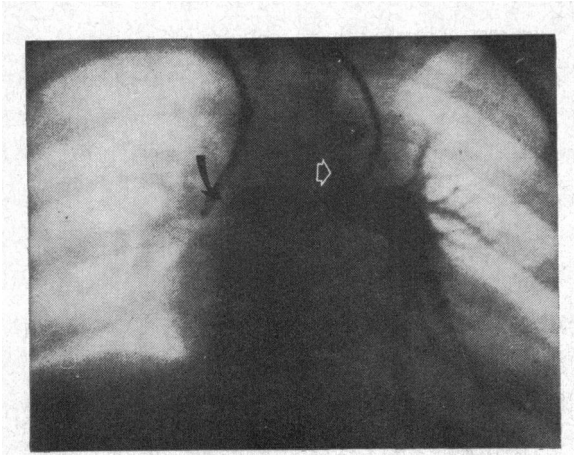


FIG. 1 *Case 2. Pulmonary blood supply from a persistent ductus arteriosus. A transarterial catheter has traversed the persistent ductus arteriosus and a hand injection of contrast medium opacifies both central pulmonary arteries. A jet* (hollow white arrow) *of contrast medium runs retrogradely into the aorta. Contrast medium is washed away from the distal right pulmonary artery by a surgical aorta/right pulmonary artery anastomosis* (black arrow).

through technique (Kappagoda and Linden, 1972). In 3 patients (Cases 2, 3, and 5) studied before the development of that method, oxygen uptake was predicted from the equations of Kappagoda *et al.* (1973). Pulmonary (Qp) and systemic (Qs) blood flows were determined by the Fick method, with mixed venous oxygen saturation taken to equal superior vena caval saturation, and pulmonary venous saturation assumed to be 97 per cent with a Po_2 of 100 mmHg.

In the 2 patients (Cases 6 and 7) with major aorto-pulmonary arteries alone supplying the intrapericardial pulmonary arteries, the tip of the catheter was advanced to the hilum and its position demonstrated by a hand injection of contrast medium. If blood could be freely sampled from that site, and contrast medium injected as a slow trickle could be seen to opacify retrogradely the intrapericardial pulmonary arteries, the pressure at that site was taken to be the maximum possible sixth aortic arch pressure (Pp6). In all patients Rpao was calculated as mean aortic pressure ÷ Qp, and where appropriate (see Discussion) Rp6 was calculated as mean sixth aortic arch pressure ÷ Qp.

When the catheter was withdrawn from the pulmonary artery to the aorta, the site of any pressure gradient was recorded by a single *x*-ray film of the catheter at that site, so that this could be compared with the other angiocardiograms.

In 2 patients (Cases 7 and 8) with pulmonary blood supply from collateral arteries, because surgical ligation of these arteries was contemplated, the haemodynamic effects of this intervention were tested by trial balloon occlusion. A No. 6 Edwards balloon catheter was advanced from an axillary arteriotomy into the descending aorta, and its tip advanced as far as possible into the largest collateral artery observed. The balloon was then inflated until it occluded the collateral artery. The femoral artery pressure was monitored throughout inflation, and remained unchanged, thus demonstrating

Qp (*l./min per m²*)	*Qs* (*l./min per m²*)	*Qp/Qs*	*Rpao units m²*	*Rp6 units m²*	*Rs units m²*	*Post bypass pressure* (*mmHg*)	
1·3	—	—	—	3·0	—		
1·6	5·8	0·3	46·2	6·4	12·9		
3·5	3·5	1·0	28·4	4·3	28·4	RV 25/4 LV 95/5	
2·9	2·7	1·1	28·6	1·4	30·7		
5·8	4·3	1·3	13·1	11·7	19·8	RV 45/5 Ao 105/80	Left pulm. artery pressure measured at operation.
10·1	5·4	1·9	8·7	6·4	16·3		After 100% O_2 for 10 min.
3·6	3·9	0·9	28·3	> 7·8	19·4		Left pulm. artery pressure measured at operation.
5·9	2·0	2·0	10·6	> 3·9	24·3		
3·6	5·6	0·6	21·6	> 12·5	13·0		
6·3	2·6	2·4	11·1	9·2	26·9		Main pulm. artery pressure measured after 100% O_2 for 10 min.

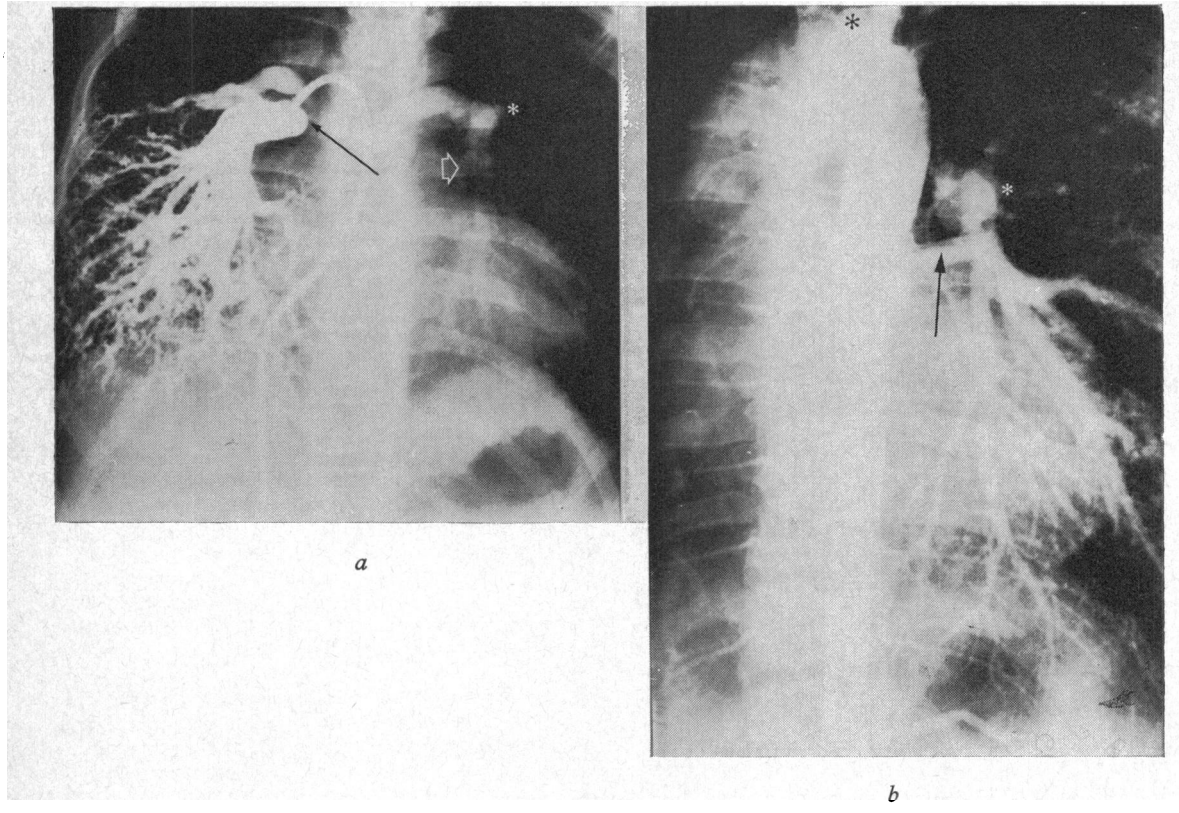


FIG. 2 *a*) *Case 5. Right pulmonary arteriogram. A transvenous catheter has been passed through the right ventricle into the overriding aorta and across a surgical aorta/lower right pulmonary artery anastomosis. Thrombosis of the vessels to the right upper zone had occurred. There is retrograde opacification of the proximal right and left pulmonary arteries, and a stenosis* (black arrow) *of the lower right pulmonary artery at the site of the anastomosis. The white arrow shows the site of entry of the collateral vessel demonstrated in Fig. 2b. Flow from this vessel causes dilution of contrast medium in the descending left pulmonary arteries. b*) *Same patient. Descending aortogram. A short collateral artery* (black arrow) *links the descending aorta with the descending left pulmonary artery. To help comparison with Fig. 2a a white asterisk has been placed in an identical site in the two pictures. Analysis of this and subsequent frames demonstrated that vessels opacified in the left lung on both these injections were identical. A black asterisk marks the stump of a thrombosed aortic arch collateral.*

that no compromise of the aortic lumen had occurred. The balloon was then deflated, and after a five-minute rest period, control measurements of systemic flow (Qs) and resistance (Rs) were made. Since it had been established that systemic and pulmonary arterial saturations were identical, Qp could be calculated without a catheter in the pulmonary artery. The balloon was then reinflated, and the measurements repeated until they had stabilized. After 15 to 20 minutes, the balloon was deflated, and control measurements repeated.

Complete occlusion of the aortopulmonary collateral artery was taken to have occurred when the balloon took on the shape of the collateral artery (Fig. 3), the pressure measured through the central lumen of the catheter (i.e. distal to the balloon) became non-phasic, and a hand injection of contrast medium down the central lumen of the catheter remained stagnant in the occluded artery.

Results

The systemic arterial and pulmonary arterial oxygen saturations were not significantly different in any patient except Case 1, where partial occlusion of the ductus resulted in unstable conditions.

On the basis of angiocardiography, three groups of patients were recognized, whose haemodynamic data are summarized in the Table.

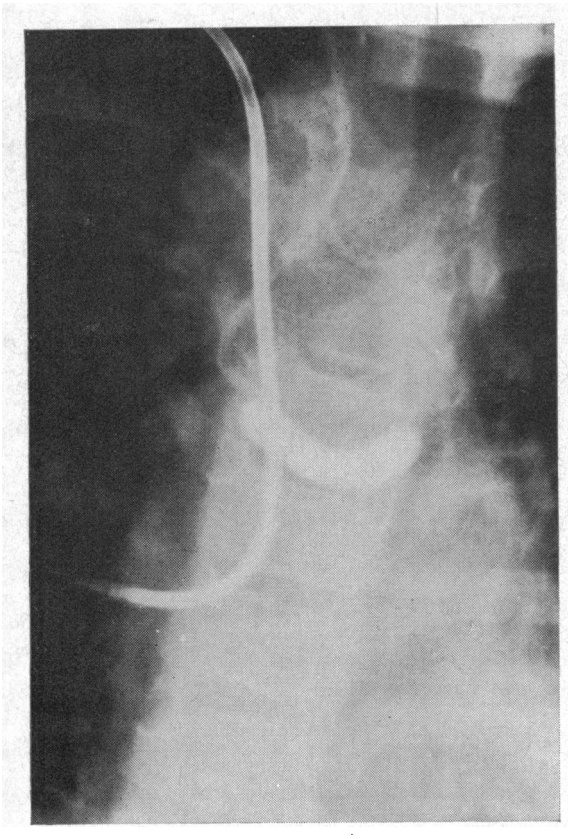


FIG. 3 *Balloon occlusion of major aortopulmonary collateral (Case 8). The balloon catheter has been passed down a right-sided descending aorta into the collateral artery. On inflation with contrast medium it takes up the shape of the artery (cf. Fig. 6b). Another catheter is in the right atrium for taking mixed venous samples.*

Group 1 (4 patients): pulmonary blood supply from persistent ductus arteriosus: no major aortopulmonary collateral arteries

Three of these patients, by the time of the investigation described, had had prior palliative surgery. Two had had an anastomosis between the ascending aorta and right pulmonary artery, and one had had a right subclavian/pulmonary artery anastomosis. In the latter, the originally persistent ductus arteriosus had closed subsequent to palliative surgery. In the remainder the ductus was persistent.

In patients with both a ductus arteriosus and an ascending aorta/right pulmonary artery anastomosis, aortography did not help in demonstrating whether both were persistent or only one, and if so which. Even if patency of the ductus could be demonstrated by a carefully sited injection well distal to the surgical anastomosis or into the ductus itself, analysis of a subsequent aortic root injection was not found to help in deciding whether the surgical anastomosis was also patent. Passage of the catheter through either the ductus or the anastomosis was the best method of proving patency, particularly when combined with direct injection of contrast medium into the pulmonary artery (Fig. 1).

In all but one patient (Case 4), both pulmonary arteries were entered. The pressure and oxygen saturation in each was identical. The aortic/pulmonary artery systolic pressure gradient ranged from 71 to 92 mmHg (mean 83) and the site of this pressure gradient was at the pulmonary end of both the Blalock anastomosis and the persistent ducti. In the one persistent ductus arteriosus not traversed with a catheter (Case 4), the pressure in the ductal lumen was at systemic level, but selective ductal angiography showed a severe stenosis at the pulmonary end of the ductus. Rp6 ranged from 1·4 to 6·4 units m^2 (mean 3·8).

Case 3 had had corrective surgery under cardiopulmonary bypass, with the right ventricular pressure falling to normal. The remainder will have complete correction in due course.

Group 2 (3 patients): pulmonary blood supply from major aortopulmonary collateral arteries

The anatomical basis of pulmonary arterial blood supply is pictured in part in Fig. 2, 4, and 5. In each case the total picture was built up from analysis of numerous injections of contrast medium in different sites.

In Case 5, no pulmonary arteries could be entered from the 3 aortopulmonary collateral arteries. One of these, arising from the aortic arch was angiographically shown to have thrombosed in between two cardiac catheterizations, one before palliative surgery and one before corrective surgery. All 3 arteries anastomosed with the sixth aortic arch. The right pulmonary artery was entered from the ascending aorta/right pulmonary artery anastomosis, and the mean pressure therein was only 8 mm below systemic (systolic pressure gradient 28 mmHg). The left pulmonary artery could not be entered. Had the two pulmonary artery pressures been equal, Rp6 would have been considerably raised at 11·7 units m^2. However, right pulmonary angiography indicated a stenosis of the right pulmonary artery immediately to the left of the anastomosis (Fig. 2a). At operation, before bypass, the left pulmonary artery pressure was found to be 18/5 mmHg. A Y-shaped homograft was inserted between the right ventricle and the two pulmonary arteries separately. The immediate post-bypass

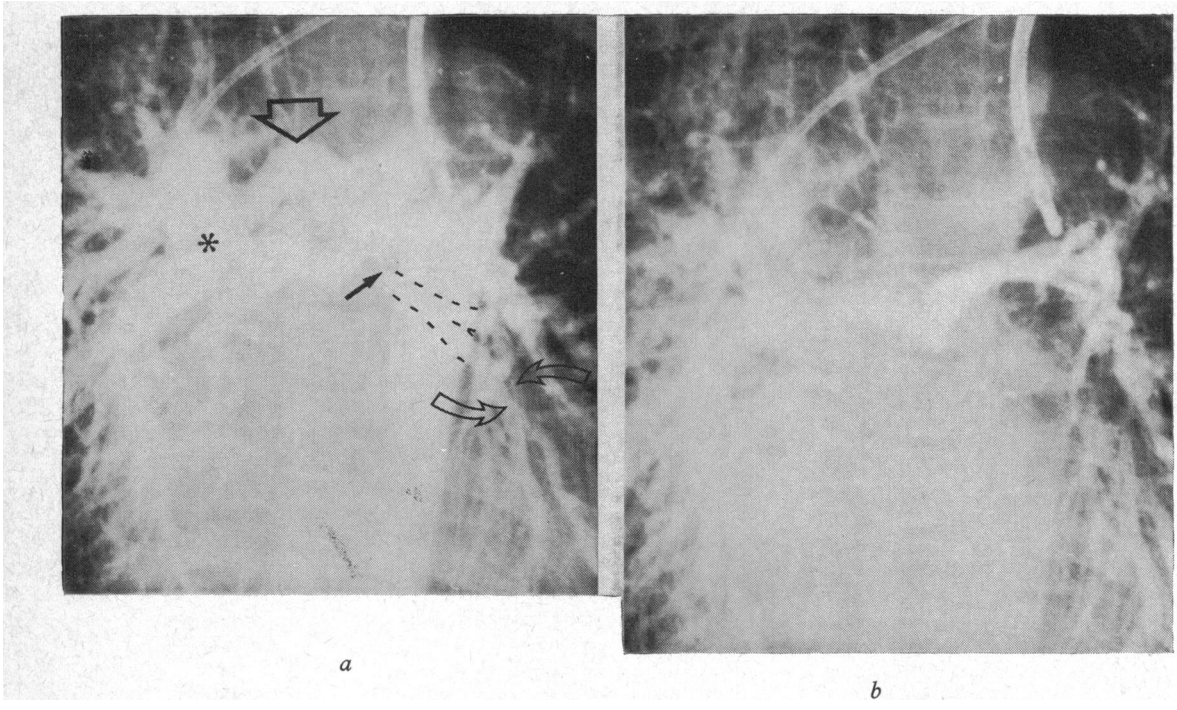


FIG. 4 *a) Case 7. Descending aortogram. Pulmonary blood supply is principally derived from a single large collateral artery from the descending aorta. A large black arrow marks the point at which this was occluded by a balloon, and an asterisk the point at which sixth aortic arch pressure was recorded. Immediately proximal to this a haemodynamic stenosis* (not visualized angiographically) *was detected. A small branch of the major collateral artery* (reinforced with dashes) *supplies part of the left lung and has a stenosis* (solid black arrow) *near its origin. The sixth aortic arches are identifiable by comparison with Fig. 4b. The curved black arrows show peripheral arteries supplied by this collateral. b) Same patient. Descending aortogram, 0·67 sec after Fig. 4a. The Y-shaped sixth aortic arches are well seen, and non-opacified blood has not only cleared the major collateral vessels, but also the proximal portions of the peripheral pulmonary arteries supplied by the left collateral artery demonstrated in Fig. 4a, suggesting that their blood supply is separate from that derived via the sixth aortic arches.*

pressure was 45/5 (RV) and 105/80 mmHg (aorta). No attempt was made to divide collateral arteries.

In Cases 6 and 7, the right pulmonary artery pressure was measured by catheterization via one aortopulmonary collateral artery, and found to be 35/24 and 34/17 mmHg, respectively. This pressure corresponded well in Case 7 with that measured in the right pulmonary artery at operation. In Case 6 the intraoperative left pulmonary artery pressure was rather lower (10/0 mmHg). In both patients the stenosis was at the distal, pulmonary end of the collateral artery. Furthermore, angiocardiography suggested the presence of at least one collateral artery which did not anastomose with the sixth aortic arch, but provided separate blood supply to a small volume of lung (Fig. 4 and 5).

Both these patients have had unilateral subclavian/pulmonary anastomoses created, with ligation of collateral arteries.

Group 3 (1 patient): mixed pulmonary blood supply

The anatomical basis of pulmonary blood supply in this patient is indicated in Fig. 6. The main pulmonary artery was supplied in retrograde fashion by a large vessel originating just proximal to the first brachiocephalic trunk (a left innominate artery in association with a right aortic arch) (Fig. 6a, c, d). This vessel is interpreted (v.i.) as a persistent fifth aortic arch.

The main pulmonary artery was entered via the fifth aortic arch and there was a 30 mmHg systolic

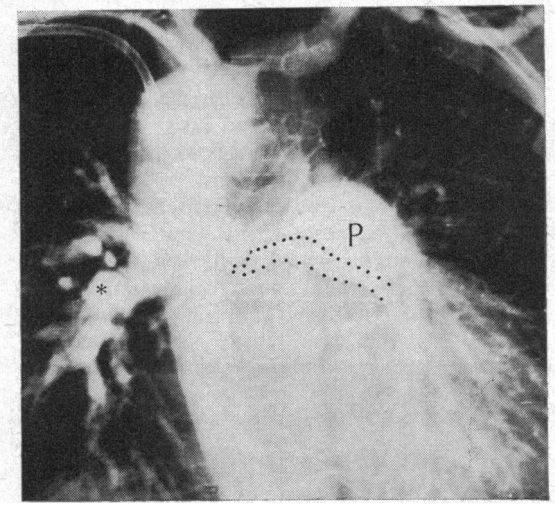


FIG. 5 *Case 6. Descending aortogram. The major pulmonary blood supply is from a large collateral artery from the descending aorta. Sixth aortic arch pressure was measured at a point (*) immediately distal to a stenosis not visualized angiographically. Note the diminution in calibre of the aorta immediately distal to the collateral. The main pulmonary artery (P) is surprisingly large and opacifies retrogradely. A separate collateral* (reinforced) *appears to supply blood to the left lung in parallel with the sixth aortic arch.*

gradient at the pulmonary/fifth aortic arch junction (Fig. 6a). Some pulmonary blood supply was also derived from collateral arteries from the descending aorta. When the catheter was advanced to a point in a left collateral artery (Fig. 6b) at the left hilum a systolic pressure gradient of 48 mmHg was encountered. Blood could be sampled with ease distal to this stenosis. Thus the intrapericardial pulmonary artery pressure was higher than that in the distal collateral artery. Not surprisingly, injection of contrast medium into the distal collateral artery failed to opacify the central pulmonary artery. This suggested that there was no functional, and perhaps no anatomical, connexion between the intrapericardial pulmonary arteries and the collateral artery. Support for this hypothesis was achieved by subtraction of the aortogram made by injection into the descending aorta from that made by injection into the apparently parallel vascular beds supplying both lungs, one fed by descending aortic collaterals and the other by the fifth aortic arch.

Balloon occlusion of aortopulmonary collateral artery The results in Case 8 are shown in Fig. 7. Occlusion caused a progressive fall in systemic arterial oxygen saturation (total 12%) over 10 minutes, after which this stabilized. At the same time pulmonary blood flow fell by 1·9 l./min per m^2 (34% of control), and systemic blood flow rose by 2·3 l./min per m^2. Systemic resistance fell by 3·7 units m^2. but mean systemic pressure rose by 6 mmHg. There was a transitory rise in systemic flow and fall in systemic resistance at 15 minutes, but the figures at 10 and 20 minutes were much the same. Five minutes after deflation of the balloon, the control state had been reached.

The results in Case 7 were similar except that systemic resistance remained unchanged during occlusion, perhaps because systemic arterial oxygen saturation fell only by 6 per cent. 660 ml/min per m^2 were transferred from the pulmonary to systemic circulation, raising mean systemic pressure by 8 mmHg.

Neither patient felt any effects of occlusion of the collateral artery.

Discussion

Consideration of the patients described in the paper, and those previously described in whom there was absence of the sixth aortic arch (Macartney *et al.*, 1973) suggests that four basic questions need to be asked about pulmonary blood supply in every patient with pulmonary atresia with ventricular septal defect.

i) Is either sixth aortic arch present?
ii) What are the sources of pulmonary blood supply?
iii) How do these sources of blood supply connect with (a) the sixth aortic arch, if present, and (b) the peripheral pulmonary arteries?
iv) What resistance to blood flow will be offered by the network of pulmonary vessels linked to the right ventricle by a conduit at the time of 'corrective' surgery?

i) Identification of sixth aortic arches

Radiographically, these appear in the expected sites of the left and right pulmonary arteries and the ductus arteriosus. In all the cases described above there was a main pulmonary artery stub present, and the sixth aortic arches were hypoplastic. The combination of these factors produced a very characteristic Y-shaped vessel running across the mediastinum immediately posterior to the aorta (Fig. 2a, 4a, 5, and 6a).

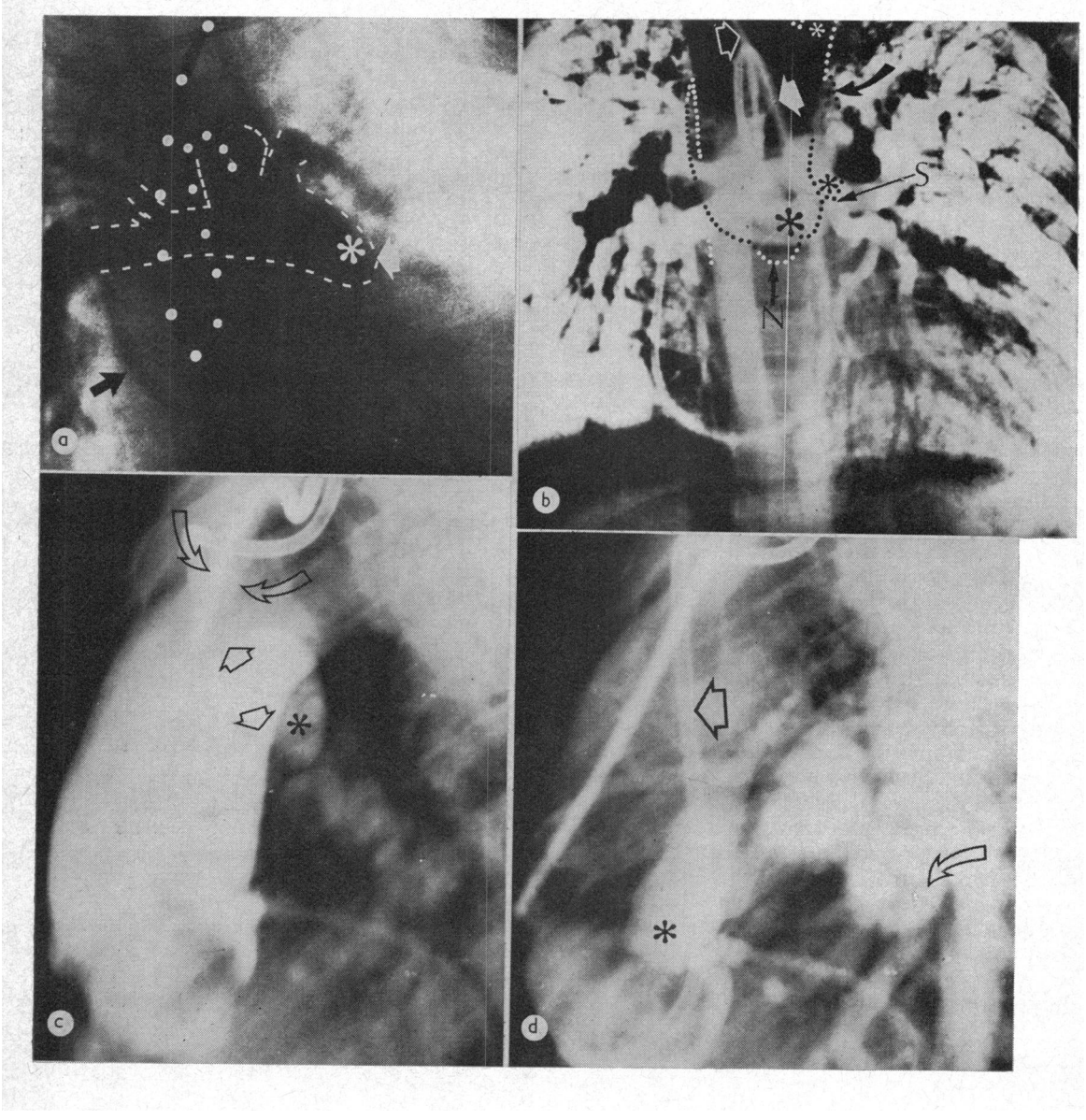
This was visualized in all cases by aortography alone. However, in one case seen by us (but not described because of failure to determine sixth aortic arch pressure), the sixth aortic arch was not

visualized on aortography, but was seen after selective injection of contrast medium into a major aortopulmonary collateral artery. Its presence was confirmed at operation. This experience confirms our view that selective catheterization of major aortopulmonary collateral arteries is an essential part of preoperative evaluation.

ii) Sources of pulmonary blood supply

There appear to be three congenital sources of pulmonary blood supply in pulmonary atresia: a) major aortopulmonary collateral arteries; b) the dorsal sixth aortic arch (persistent ductus arteriosus); and c) persistent fifth aortic arch.

The first two of these in our series were mutually exclusive. However, Jefferson, Rees, and Somerville (1972) have recorded coexistence of a persistent ductus arteriosus and major aortopulmonary collateral arteries in the same patient. The same authors report 12 patients in whom pulmonary arterial blood supply was derived from small, multiple, systemic arteries. Though this pattern of blood



supply is familiar to us in older patients with complex pulmonary atresia and tetralogy of Fallot, it has not been encountered in any of the patients studied here. We have been unable to find any published report of this kind of blood supply in an infant, and conclude that it is probably acquired and not congenital.

The existence of a fifth aortic arch in man has been disputed by embryologists for many years. However, Van Praagh and Van Praagh (1969) presented a necropsied case of duplicated left aortic arch and argued cogently that the more caudal of the two arches was a persistent fifth aortic arch. This ran from a point just proximal to the innominate artery to the aortic end of the ligamentum arteriosum.

The vessel in Case 8 which supplied the main pulmonary arteries occupies a site which is in fact even more typical of the fifth aortic arch. Congdon (1922) states that of the 6 well-developed so-called fifth aortic arches that have been described in the human embryo, 5 *enter the pulmonary arch* near its

FIG. 6 *a*) *Case 8. A Sones catheter* (solid white circles) *has been looped in the aortic root and advanced into the persistent left fifth aortic arch. A hand injection of contrast medium in this site outlines the left fifth and both sixth aortic arches* (reinforced with white dashes). *The atretic pulmonary valve is marked by a white arrow. A black arrow shows a transvenous catheter in the right atrium. The white asterisk denotes the site of measurement of pulmonary artery pressure. Note how the peripheral lung vessels opacified from the sixth aortic arch correspond with those outlined in black in Fig. 6b. b) Same patient. A composite subtraction frontal plane aortogram made by superimposing a positive film made during injection of contrast medium into the ascending aorta upon a negative film made during injection into the descending aorta. Because of a slight difference in patient position during the two injections each half of the picture has been subtracted separately. The film pair was chosen because of comparable opacification of the lung fields and descending aorta in each. Any vessel outlined in black has, therefore, opacified from injection into the ascending aorta alone, whereas vessels opacified on both injections (e.g. the descending aorta) are outlined in pale grey. The injection into the ascending aorta was made via a catheter (not visualized) passed retrograde down the left innominate artery* (white asterisk). *The injection into the descending aorta was made through a transvenous catheter* (solid white arrow) *which had been advanced from the right ventricle across the overriding aortic valve into the ascending aorta and round the right-sided aortic arch.*

There is a leash of tortuous collateral vessels arising from the descending aorta outlined in pale grey, one of which was occluded by a balloon catheter subsequently (large black asterisk, *c.f. Fig. 3*) *and had a stenosis (S) not visualized radiographically distal to which the pressure was significantly lower than sixth aortic arch pressure. Because these collateral vessels in the mediastinum largely overlap the aortic root in this projection, they obscure its outline after subtraction, except for the non-coronary aortic sinus (N). To clarify the anatomy, the course of the ascending aorta and upper descending aorta has been outlined with black or white dots as appropriate. Also seen is the fifth aortic arch* (curved black arrow) *and the main pulmonary artery* (small black asterisk). *No attempt has been made to outline the right pulmonary artery, which is also obscured by collaterals (but see Fig. 6a). However it is evident that the peripheral lung vessels outlined in black can only have been opacified via the sixth aortic arches, since these in turn have opacified from the ascending aorta via the fifth aortic arch (c.f. Fig. 6a, c, and d). The vessels in black have not opacified from collaterals from the descending aorta, which therefore provide a source of pulmonary blood flow which is independent of the sixth aortic arches.*

The hollow white arrow points to a catheter lying in the ascending aorta during injection into the descending aorta. c) Same patient. Lateral projection of aortic root angiogram. The transarterial catheter used for injection has traversed the left innominate artery (origin shown by curved arrows). *There is a right aortic arch (Fig. 6b) so this is the first brachiocephalic trunk, derived from the embryological left fourth aortic arch. Proximal to this* (straight arrows) *is the origin of a large vessel* (asterisk), *the persistent left fifth arch. d) 0·17 sec after Fig. 6c, retrograde opacification of the main pulmonary artery* (*) *has occurred (cf Fig. 6a). Though the great arteries appear transposed, analysis of the right ventricular angiogram showed mitral/aortic continuity (i.e. origin of aorta from left ventricle). Clearing of contrast medium from the ascending aorta has exposed a transvenous catheter* (straight arrow). *Opacification of collateral arteries from the descending aorta* (curved arrow) *has also occurred.*

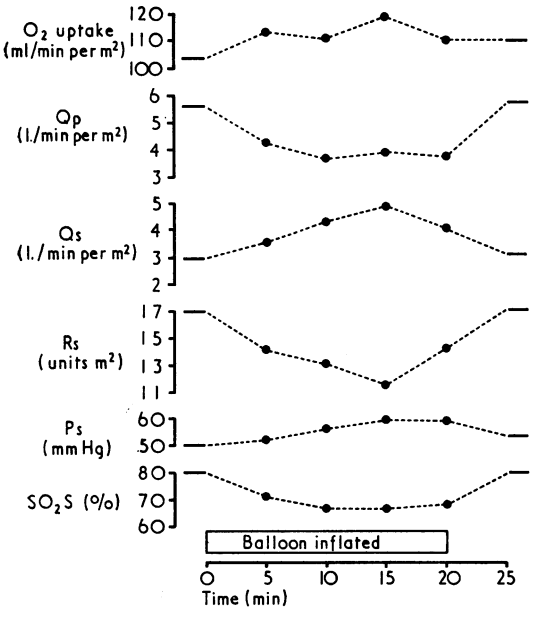


FIG. 7 *Effect of balloon occlusion of aortopulmonary collateral artery (Case 8). For explanation see text. Qp = pulmonary blood flow. Qs = systemic blood flow. Rs = systemic resistance. Ps = systemic arterial mean pressure. SO_2S = systemic arterial oxygen saturation.*

termination (our italics). As the angiocardiograms clearly demonstrate (Fig. 6), this vessel originated proximal to the left fourth arch (the left innominate artery) and ran between it and the left ventral sixth arch (the proximal left pulmonary artery), entering the latter at precisely the point where it would normally continue into its dorsal portion (the ductus arteriosus).

Establishing whether or not the ductus arteriosus is patent is important for two reasons. Firstly, it provides access to the otherwise rather inaccessible left pulmonary artery. Secondly, it must be closed, if patent, at the time of complete correction. Catheterization of the ductus by the method described is the most reliable way of determining patency.

iii) Connexions with sixth aortic arch and peripheral pulmonary arteries

In each patient studied, the congenital source of pulmonary blood supply, the sixth aortic arch, and the peripheral pulmonary arteries anastomosed with one another at one or both hila. Furthermore, it was at the site of the systemic/sixth aortic arch anastomoses that stenosis tended to occur. Systolic pressure gradients of 60 mmHg and more were located in the absence of any angiographically demonstrated stenosis (Fig. 4, 5, and 6b). Therefore angiocardiography alone is unreliable in assessing the haemodynamic state. Because of these stenoses sixth aortic arch pressure was invariably less than systemic arterial pressure.

What is of equal importance, however, is that the sixth aortic arches did not necessarily connect with all peripheral pulmonary arteries. In Cases 6, 7, and 8 at least one aortopulmonary collateral artery appeared to supply a section of lung not supplied via the sixth aortic arch. This situation has previously been reported in one patient studied both in life and at necropsy (Jefferson *et al.*, 1972 – Case 13). In such patients pulmonary blood supply is *multifocal*, in that there is no common 'focus' for pulmonary blood supply other than the aorta. By contrast, the patients in group 1, and Case 5 had *unifocal* pulmonary blood supply, in that the sixth aortic arches provided an additional common 'focus' (or pathway) for all blood flow to the lungs. Fig. 8 demonstrates this distinction diagrammatically.

iv) Measurement of pulmonary resistance

To obtain a meaningful measurement of pulmonary resistance in pulmonary atresia by the Fick principle, the oxygen saturations and blood pressures in all vessels supplying the lungs must be essentially the same, and the reference point from which a pulmonary resistance is measured must be defined (Macartney *et al.*, 1973). To be of practical value this reference point must also be the point to which a surgical conduit from the right ventricle is connected.

All these criteria are fulfilled if pulmonary resistance relative to the aorta (Rpao) is calculated, but the resistance thus measured was $> 10{\cdot}7$ units m^2 in all patients. However, this high resistance is at least in part due to stenoses at the junctions between systemic arteries and the sixth aortic arch, whether congenital or surgical. These may be largely bypassed if the surgical conduit is anastomosed to the sixth aortic arch. Under these circumstances, however, pulmonary resistance relative to the sixth aortic arch pressure (Rp6) can only be calculated if pulmonary blood supply is unifocal and the pressure in each pulmonary artery is more or less equal. On this basis all patients in group 1 are regarded as suitable for total correction.

Where pulmonary blood supply is multifocal no measurement of Rp6 can be made if sixth aortic arch pressure is known, unless the blood flow (Qp6) through that portion of lung supplied by the sixth aortic arch can be measured. Since selective injections of contrast medium into aortopulmonary

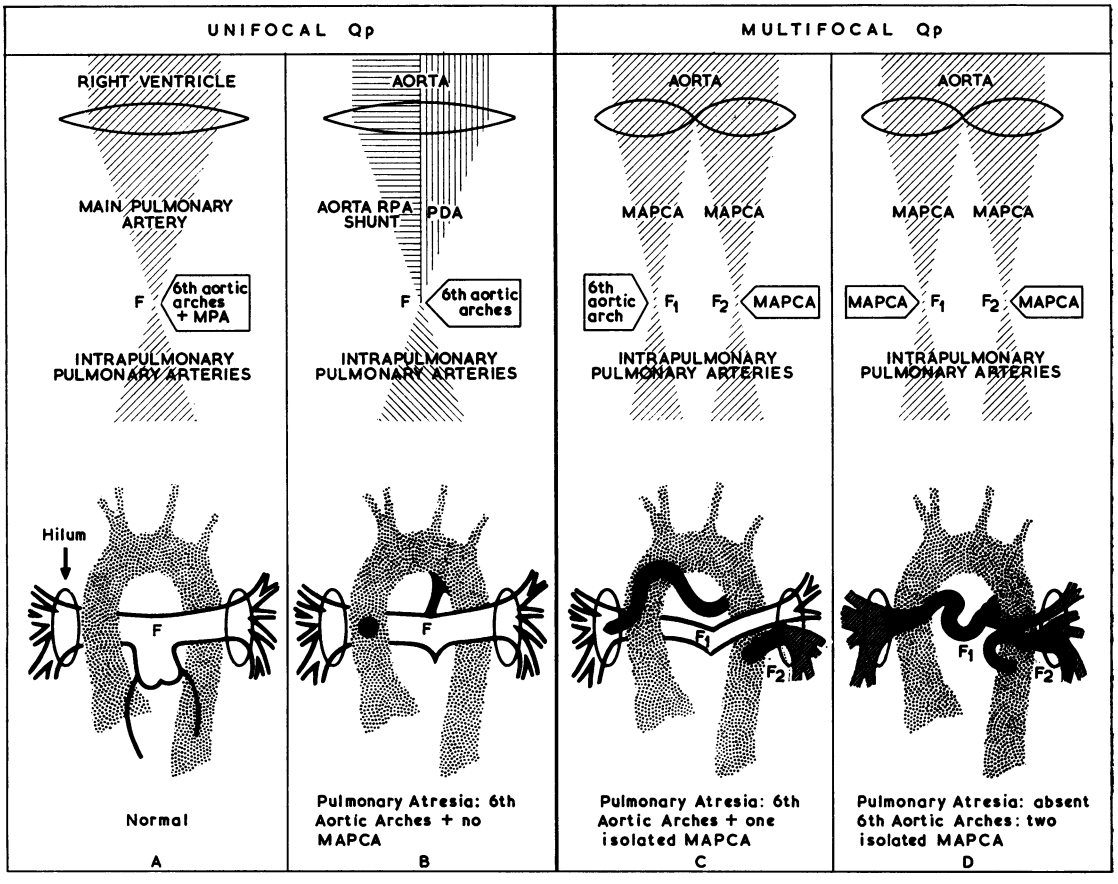


FIG. 8 *Diagrammatic illustration of the distinction between unifocal and multifocal pulmonary blood flow (Qp). In each panel (A, B, C, D) the anatomical situation in the lower picture is interpreted in the upper diagram in terms of a lens focusing parallel light rays. A focus* (see *F*, F_1, F_2) *is defined as any isolated vessel or complex of vessels lying between the 'original source' of pulmonary blood supply and the intrapulmonary arteries which provides pulmonary blood supply at an essentially common pressure throughout the extent of the focus. Thus, in the normal individual (panel A) the 'original source' is the right ventricle and the main pulmonary artery and sixth aortic arches form a single focus (F). Panel A also illustrates the desired result of corrective surgery. In pulmonary atresia with VSD (panels B, C, D), the aorta is the 'original source' of pulmonary blood supply. Aortopulmonary anastomoses are drawn in solid black. Vessels supplying independent portions of lung but not anastomosing with either sixth aortic arch are striped, and their presence indicates that multifocal blood supply is present.*

In panel B, all vessels supplying the lungs anastomose with the sixth aortic arch. Thus, though there are two *aortopulmonary anastomoses (surgical aorta – right pulmonary artery (RPA) shunt and persistent ductus arteriosus (PDA)) there is only* one *focus (the sixth aortic arches).*

In panel C, there are two aortopulmonary anastomoses (i.e. major aorto-pulmonary collateral arteries (MAPCA)). One anastomoses with the right sixth aortic arch at the hilum, so the sixth aortic arch forms one focus (F_1). However, a second MAPCA provides an independent second focus (F_2) for pulmonary blood supply.

In panel D, there is no sixth aortic arch, and there are two MAPCA forming independent foci. The location of F_1 indicates that the pressure distal to the branch point of the collateral is the same in both branches as far as the hilum. Absence of the sixth aortic arch is (Macartney et al., *1973) not discussed in this paper but included in the diagram to show its relation with other types of pulmonary atresia.*

The cases illustrated by no means exhaust the possible interrelations of aorta, anastomoses, and intrapulmonary pulmonary arteries. It is, for example, conceivable that unifocal pulmonary blood supply can exist in absence of the sixth aortic arch (e.g. Cases 1 and 2, Macartney et al., *1973).*

collateral arteries not anastomosing with the sixth aortic arch show a normal pulmonary capillary blush and pulmonary venous filling, it must be presumed that they participate in oxygen uptake. Qp6 cannot, therefore, be measured by the classical Fick method. A minimum value of Rp6 can be calculated on the theoretical assumption that Qp6 = total Qp. In Case 8, minimum Rp6 was 12·5 units m^2, implying severe pulmonary vascular disease in that region of lung supplied by the sixth aortic arch. Therefore, demonstration of central pulmonary arteries is no guarantee of operability in pulmonary atresia. The surgical approach of a preliminary operation designed to ligate major aortopulmonary collateral arteries and substitute surgical aortopulmonary anastomoses is suggested for two reasons. The first concerns the problem of ligation of anastomoses at the time of total correction. Major aortopulmonary collateral arteries enter the hilum from behind, so access to them is extremely difficult if a mid-line sternotomy is used to expose the heart for intracardiac correction. Surgical anastomoses are much more accessible from this approach.

The second advantage of preliminary surgical operation is that if all collaterals, and in particular those that do not anastomose with the sixth aortic arch, be ligated then pulmonary blood supply becomes unifocal, and the Rp6 can be determined with confidence upon recatheterization. The technique of balloon occlusion of aortopulmonary collateral arteries described may make such an operation unnecessary in certain circumstances. If pulmonary blood supply were unifocal apart from one major aortopulmonary artery, temporary occlusion of that artery would enable Rp6 to be calculated by determination of total Qp and Pp6 during occlusion. That collateral, together with the others, could then be divided at the time of complete correction by the method of Doty *et al.* (1972).

If direct measurement of Qp6 at the time of cardiac catheterization were feasible in all cases of multifocal pulmonary blood supply, this second advantage would exist no longer. We are currently exploring methods of determining Qp6 in these circumstances.

Grateful thanks are expressed to Dr. P. G. Keates for preparing the subtraction films for Fig. 6b, and to Miss Jane Artle for technical help in preparing the manuscript.

References

Chesler, E., Beck, W., and Schrire, V. (1970). Selective catheterization of pulmonary or bronchial arteries in the preoperative assessment of pseudotruncus arteriosus and truncus arteriosus type IV. *American Journal of Cardiology*, **26**, 20.

Congdon, E. D. (1922). Transformation of the aortic-arch system during the development of the human embryo. *Contributions to Embryology (Publications of the Carnegie Institution)*, **14**, 47.

Doty, D. B., Kouchoukos, N. T., Kirklin, J. W., Barcia, A., and Bargeron, L. M., Jr. (1972). Surgery for pseudotruncus arteriosus with pulmonary blood flow originating from the upper descending thoracic aorta. *Circulation*, **45**, Suppl. I, 121.

Edwards, J. E., and McGoon, D. C. (1973). Absence of anatomic origin from heart of pulmonary arterial supply. *Circulation*, **47**, 393.

Jefferson, K., Rees, S., and Somerville, J. (1972). Systemic arterial supply to the lungs in pulmonary atresia and its relation to pulmonary artery development. *British Heart Journal*, **34**, 418.

Kappagoda, C. T., Greenwood, P., Macartney, F. J., and Linden, R. J. (1973). Oxygen consumption in children with congenital diseases of the heart. *Clinical Science and Molecular Medicine*, **45**, 107.

Kappagoda, C. T., and Linden, R. J. (1972). A critical assessment of an open circuit technique for measuring oxygen consumption. *Cardiovascular Research*, **6**, 589.

Macartney, F., Deverall, P. B., and Scott, O. (1973). Haemodynamic characteristics of systemic arterial blood supply to the lungs. *British Heart Journal*, **35**, 28.

McGoon, D. C., Rastelli, G. C., and Ongley, P. A. (1968). An operation for the correction of truncus arteriosus. *Journal of the American Medical Association*, **205**, 69.

Miller, G. A. H., Restifo, M., Shinebourne, E. A., Paneth, M., Joseph, M. C., Lennox, S. C., and Kerr, I. H. (1973). Pulmonary atresia with intact ventricular septum and critical pulmonary stenosis presenting in first month of life. Investigation and surgical results. *British Heart Journal*, **35**, 9.

Ross, D. N., and Somerville, J. (1966). Correction of pulmonary atresia with a homograft aortic valve. *Lancet*, **2**, 1446.

Rudolph, A. M., Heymann, M. A., and Spitznas, U. (1972). Hemodynamic considerations in the development of narrowing of the aorta. *American Journal of Cardiology*, **30**, 514.

Somerville, J., and Ross, D. (1972). Long-term results of complete correction with homograft reconstruction in pulmonary outflow tract atresia. *British Heart Journal*, **34**, 29.

Tandon, R., Hauck, A. J., and Nadas, A. S. (1963). Persistent truncus arteriosus. A clinical, hemodynamic, and autopsy study of nineteen cases. *Circulation*, **28**, 1050.

Van Praagh, R., and Van Praagh, S. (1969). Persistent fifth arterial arch in man. Congenital double-lumen aortic arch. *American Journal of Cardiology*, **24**, 279.

Victorica, B. E., Krovetz, L. J., Elliott, L. P., van Mierop, L. H. S., Bartley, T. D., Gessner, I. H., and Schiebler, G. L. (1969). Persistent truncus arteriosus in infancy; a study of fourteen cases. *American Heart Journal*, **77**, 13.

Requests for reprints to Dr. F. J Macartney, Leeds Regional Thoracic Surgical Centre, Killingbeck Hospital, York Road, Leeds LS14 6UQ.